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Jugulotympanic Paraganglioma: A Rare Cause of Vertigo

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Statistical Analysis C
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Patient: Female, 83
Final Diagnosis: Jugulotympanic paraganglioma
Symptoms: Dizziness • tinnitus
Medication: —
Clinical Procedure: —
Specialty: Oncology

Objective: Rare disease

Background: Jugulotympanic paraganglioma generally presents in the 5th or 6th decades of life with tinnitus and hearing loss. In this manuscript, we present a rare case of jugulotympanic paraganglioma presenting in the 9th decade with vertigo as the most bothersome symptom.

Case Report: An 83-year-old woman presented with worsening episodes of dizziness of a few months duration. She also complained of tinnitus and hearing loss, more severe on the left side. Examination revealed a red bulging left-sided tympanic membrane, conductive hearing loss, and a bruit at the base of the skull. Dix-Hallpike test was negative. CT head and MRI brain revealed findings consistent with a large left-sided jugulotympanic paraganglioma, which was found to be hormonally inactive on laboratory tests. The patient underwent treatment with radiotherapy, which resulted in partial improvement of symptoms.

Conclusions: Jugulotympanic paraganglioma may manifest in the elderly with the chief complaint of intermittent vertigo, as in our case. A red bulging mass on otoscopy raises the suspicion, necessitating further investigations, including CT and MRI.

MeSH Keywords: Glomus Jugulare Tumor • Hearing Loss, Conductive • Radiotherapy • Tinnitus • Vertigo

Full-text PDF: http://www.amjcaserep.com/abstract/index/idArt/893366

Background

Jugulotympanic paragangliomas is a rare tumor that generally presents in the 5th or 6th decades of life with pulsatile tinnitus and hearing loss. In this report, we present an unusual case of jugulotympanic paraganglioma presenting in an elderly woman in the 9th decade of life with vertigo as the most worrisome symptom.

Case Report

An 83-year-old woman presented with several weeks of worsening episodes of "dizziness", which she described as unsteadiness and a tendency to fall without a sense of rotation. Although she had the mild form of these symptoms for the past 2–3 years, she had noticed the apparent worsening for the last 4 months. These episodes were precipitated by sudden changes in head position but were not preceded by any sensation of ear fullness or ear pain. She also complained of increasing bilateral hearing loss (more on the left side), as well as intermittent tinnitus. She denied any otorrhea, nausea, palpitations, ataxia, weakness, or any recent viral illness. Her past medical history was negative for any cerebrovascular accident, migraine headaches, head and neck trauma, or radiation exposure. She denied any history of smoking and alcohol or drug abuse.

On otoscopic examination of the left ear, an erythematous bulging tympanic membrane was noted. Conductive hearing loss was noticed bilaterally on Rinne test (more on the left side). No nystagmus was noted and Dix-Hallpike test was negative. A bruit was heard at the base of the skull, grade +3 on the left side. A thorough neurological examination produced normal results, including finger-to-nose test and tandem gait. Romberg test failed to reveal any truncal ataxia. She was prescribed a trial of meclizine as needed for dizziness.

The patient was evaluated by an otorhinolaryngologist, and on otoscopy, the tympanic membrane appeared fluid-filled. Myringotomy with aspiration was attempted with the expectation of a middle ear effusion, but the palpation of a soft doughy mass led to the cessation of the procedure and the concern for either jugulare paraganglioma or tympanic paraganglioma. Computerized tomography (CT) scan of the temporal bones without contrast demonstrated a nonspecific soft-tissue mass extending from the left jugular foramen into the basilar cistern, with erosion into the hypotympanum and mesotympanum extending into the tympanic cavity. MRI of the brain showed findings consistent with a large paraganglioma on the left side that extended from just above the carotid bifurcation through the jugular foramen into the tympanic cavity. In the cervical region, maximum transverse dimensions

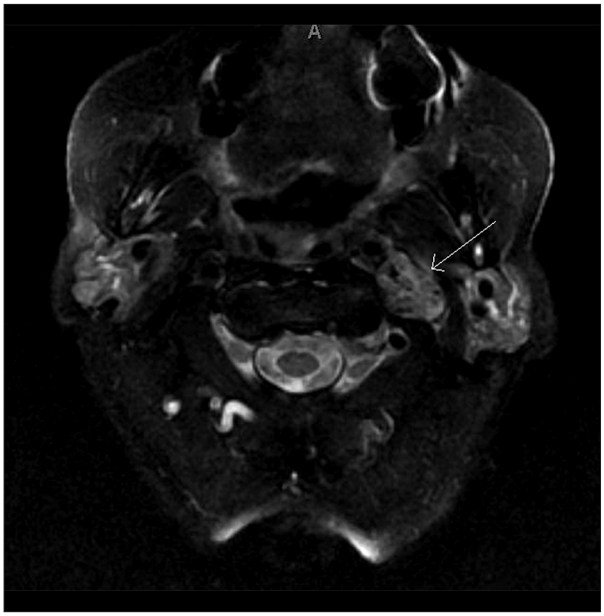

Figure 1. MRI brain with gadolinium contrast; fast spin-echo fat-suppressed T2 axial sequence demonstrating minimally hyperintense lesion in the cervical region with maximum transverse dimension of approximately 12×16 mm.

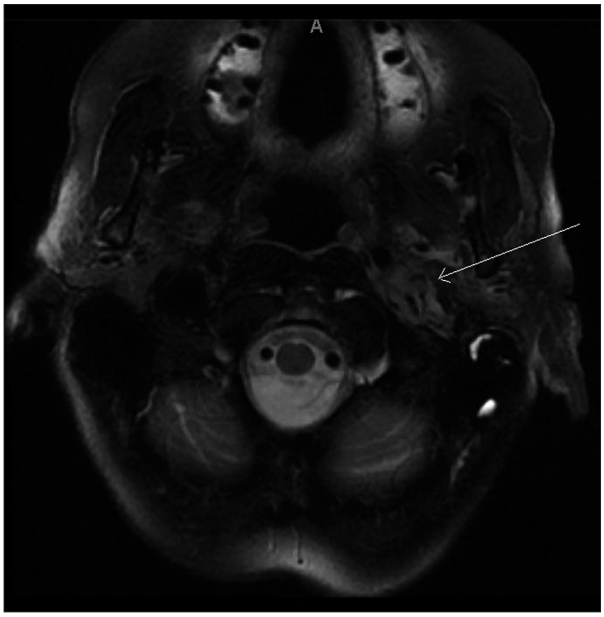

Figure 2. MRI brain with gadolinium contrast; fast spin-echo fat-suppressed T2 axial sequence demonstrating a lesion at the skull base, at jugular foramen level, dimensions approximately 26×14 mm.

were approximately 22×16 mm (white arrow in Figure 1). At the skull base, jugular foramen level, dimensions were approximately 26×14 mm (white arrow in Figure 2). In Figure 3, a T1 coronal sequence demonstrates the proximity of the

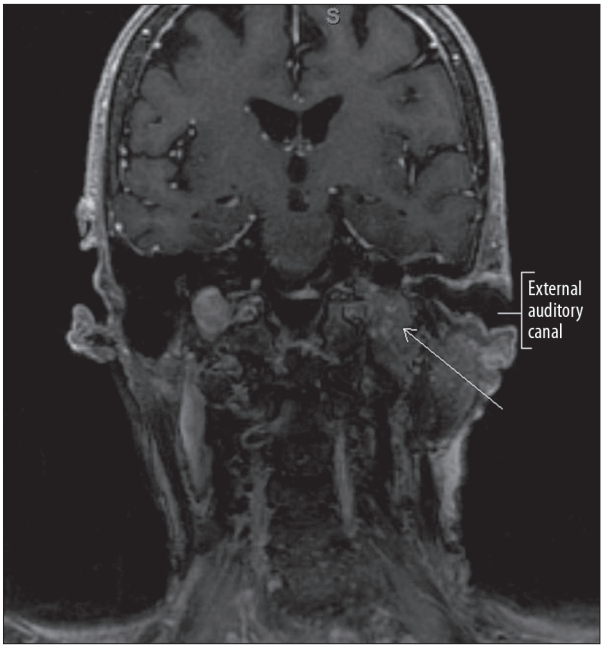

Figure 3. MRI brain with contrast; T1 coronal sequence demonstrating a mass close to the external auditory canal with diffuse minimal somewhat heterogeneous enhancement.

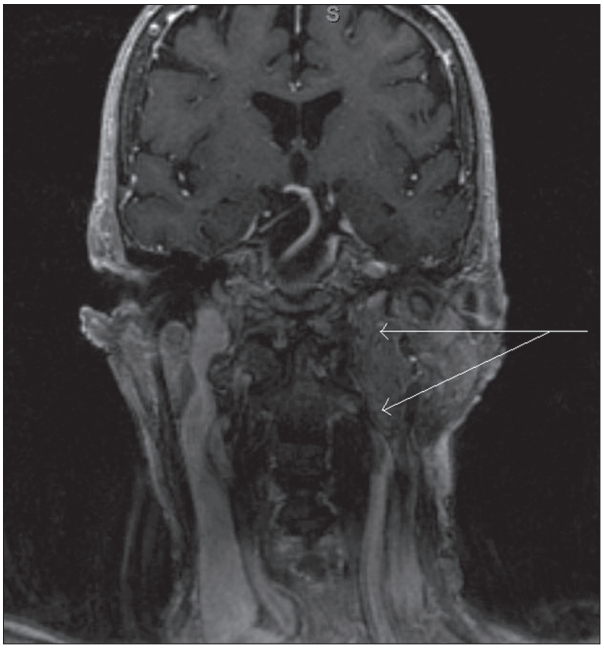

Figure 4. MRI brain with contrast; T1 coronal sequence revealing a mass in the carotid spaces with cephalocaudad dimension of approximately 50–60 mm. The caudad margin of the mass begins approximately 15–20 mm superior to the carotid bifurcation. The mass extends cephalad through the jugular foramen and left aspect of the basilar cistern with erosion into the hypotympanum and mesotympanum.

tumor to the external auditory canal. The cephalocaudad dimension was approximately 50–60 mm (white arrows in Figure 4). These findings appeared to be consistent with jugulotympanic paraganglioma. Urine epinephrine, norepinephrine, dopamine and 24-h 5-hydroxyindoleacetic acid and vanillylmandelic acid were all within normal range. She was treated with fractionated stereotactic radiotherapy, which she tolerated well. Follow-up MRIs showed partial reduction in the size of the paraganglioma. Although she had improvement in tinnitus, she continued to have significantly reduced hearing on the ipsilateral side, with unsteady gait.

Discussion

Paragangliomas are a family of benign, but locally invasive, hypervascular neoplasms that are rare, accounting for just 0.012% of all tumors [1,2]. They are found on the carotid body, the vagus nerve, along the internal jugular vein, or in the tympanic cavity. Jugulotympanic paragangliomas generally present in the 5th or 6th decades of life with pulsatile tinnitus and hearing loss. In this report, we describe an uncommon presentation of jugulotympanic paraganglioma in the 9th decade of life with vertigo as the chief complaint. Vertigo associated with tinnitus and hearing loss suggests Meniere's disease and our patient presented with a similar constellation of symptoms. However, careful examination demonstrated an erythematous bulging tympanic membrane, which prompted further investigations revealing an underlying jugulotympanic paraganglioma.

Jugulotympanic paragangliomas can present with symptoms secondary to mass effect on surrounding structures, including vasculature or the lower cranial nerves (VII, IX, X, IX) [1,3,4] like facial palsy, hearing loss (conductive or sensory), tinnitus, dysphagia, hoarseness, pain, or dizziness. Our patient denied any facial weakness, dysphagia, hoarseness, or pain; her main complaint of dizziness and associated tinnitus were likely from the mass defect of the tumor. Her hearing loss was bilateral and conductive; however, it was significantly more on the left side, which was probably contributed to by the location of the tumor. Otoscopic examination of a middle ear paraganglioma can demonstrate a reddish-blue, pulsatile mass medial to the tympanic membrane [5]. Myringotomy is contraindicated in cases of reddish mass behind the tympanic membrane, but in our patient it was performed with the expectation of middle ear effusion with the unanticipated finding of a soft mass behind the tympanic membrane. Catecholamine secretion occurs in only 2% of cases, and the tumor in our case was hormonally inactive, as well. Investigations should include head CT scans to identify the extent of bone destruction and MRI for determining tumor extension; T2-weighted images show "salt and pepper" appearance of the paraganglioma [1,6]. If the diagnosis is unclear, a diagnostic angiography can be

confirmatory, which shows the specific vascular supply of the paraganglioma [3,7].

Because of the indolent natural history of these paragangliomas, a period of observation is often appropriate [1,8]. To be selected for treatment, paragangliomas should be symptomatic or radiologically progressive [9]. With comparable or arguably improved outcomes, and also being 25–30% less expensive than neurosurgery, new irradiation modalities are the first-line treatment for most jugulotympanic paragangliomas [1,10]. The efficacy of radiotherapy is defined not by disappearance of the tumor, but by tumor control (ie., stabilization of symptoms and absence of tumor growth) [10]. The potential late toxicity of radiotherapy suggests that treatment will inevitably evolve towards chemotherapy even though there is no established role for chemotherapy currently [1,10].

Conclusions

Jugulotympanic paragangliomas are rare tumors that usually manifest in middle age with tinnitus and hearing loss. However, clinicians should be aware of unusual presentation of this tumor in elderly patients with vertigo. A red bulging tympanic membrane should trigger further investigations with a CT scan or MRI of the brain, and myringotomy should not be attempted in such cases.

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Conflict of interest

None.

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